

Case Report

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Special Vascular Intervention in Newborn with Scimitar Syndrome

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Introduction

Scimitar syndrome is a rare and complex association of congenital cardiopulmonary abnormalities with an estimated prevalence of 1 to 3 per 100,000 births [1]. Literature demonstrated that patients presenting in infancy are more likely to have severe heart failure, Aorto-pulmonary Collaterals, and a poorer prognosis than patients presenting at a later age [2-4]

Case History

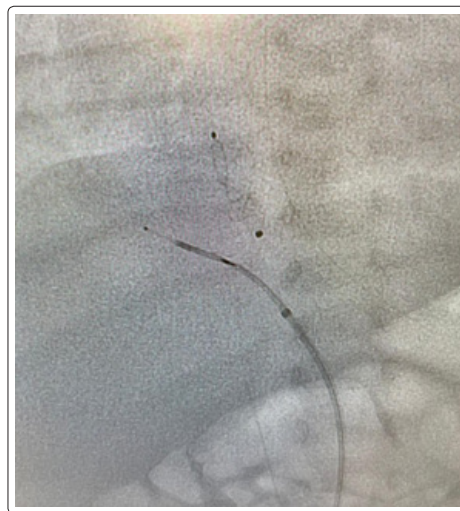
We are presenting a 4 weeks old neonate with weight of 2.7 kg, diagnosed as a non-obstructive scimitar syndrome (right lower pulmonary vein draining directly to IVC) and multiple massive aorto-pulmonary collaterals to the right lung and ASD secundum, hypoplasia of right pulmonary artery and right lung. Diagnosis was confirmed by transthoracic Echocardiography, CT scan of abdomen and thorax with contrast and cardiac MRI.

Major Aorto-Pulmonary Collaterals



Descending Aorta Angiography demonstrating multiple Major Aorto-Pulmonary Collaterals arising from Abdominal Aorta to the right lung

Deploying MVP to occlude AP collateral

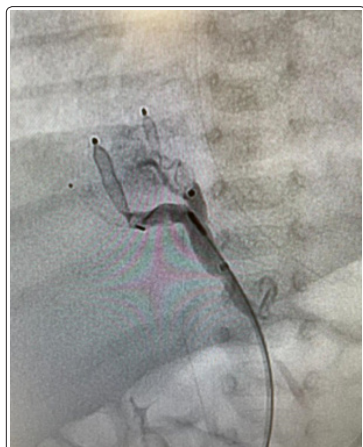


An Angiograph showing a deployed Piccolo Amplatzer occluder in one of the AP collaterals, and adjusting a microvascular plug in another AP collateral.

Case History

Patient was not diagnosed antenatally. Presented after birth with severe pulmonary hypertension requiring high setting of ventilation and high serum lactate level. Patient developed systemic circulation steal with bowel ischemia and infection (necrotizing enterocolitis). After failure of aggressive supportive medical treatment, patient was taken to the catheterization lab for diagnostic and interventional catheter.

Deploying multiple occluders for AP collaterals



An angiograph showing multiple occluders deployed in the AP collaterals



An angiograph showing multiple occluders deployed in the AP collaterals, with remaining collateral that is occluded in the last angiograph

Therapeutic Interventions

Angiographies showed multiple massive collaterals from abdominal aorta to the right lung with pulmonary venous return of the lower right lobe of the lung to the IVC and significant cardiomegaly.

We used multiple microvascular plugs, PDA occlusion devices, Piccolo device through 3 Fr then 4 Fr sheaths and catheter through right femoral artery to occlude the massive collaterals.

Final Angiographic Result



Outcome

Post- procedure patient was extubated in the NICU and heart failure symptoms disappeared.

Conclusions

Massive aortopulmonary collaterals transcatheter arterial embolization can be done safely in the catheterization lab in neonates with small body weight and represent a minimal invasive option compared to surgical option. Embolization of anomalous systemic arteries can dramatically improve symptoms of heart failure and pulmonary hypertension in neonates.

References

1. Dupuis C, Charaf LA, Breviere GM, Abou P, Remy-Jardin M, et al. (1992) The “adult” form of the scimitar syndrome. *Am J Cardiol* 70: 502-7.
2. Dusenbery SM, Geva T, Seale A (2013) Outcome predictors and implications for management of scimitar syndrome. *Am Heart J* 165: 770-7.
3. Hoffman JIE (2009) Scimitar syndrome. In: Hoffman JIE (ed). *The natural and unnatural history of congenital heart disease*. Hoboken, NJ: Wiley-Blackwell: 161-6.
4. Najm HK, Williams WG, Coles JG, Rebeyka IM, Freedom RM (1996) Scimitar syndrome: twenty years' experience and results of repair. *J Thorac Cardiovasc Surg* 112: 1161-8

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